Interrupted Inferior Vena Cava and Left-Sided Subrenal Inferior Vena Cava

Prenatal Diagnosis

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An interrupted inferior vena cava (IVC) and a left-sided subrenal (postrenal) IVC with azygos or hemiazygos continuation are very rare anomalies of the IVC. The prenatal sonographic and color Doppler features of these anomalies are reported here.

Case Reports

Case 1

A 33-year-old woman, gravida 3, para 1, aborta 1, was referred for sonography to rule out fetal anomalies. She had 1 miscarriage at 45 days’ amenorrhea and 1 live female child of 7 years. There was no history of consanguinity. Her menstrual age was 30 weeks. Sonography was done on an HDI 5000 scanner (Philips Medical Systems, Bothell, WA) with convex broadband probes of 2 to 5 and 4 to 7 MHz. Biometric measurements corresponded to the menstrual age of 30 weeks. There was an interrupted IVC with nonvisualization of the intrahepatic and suprarenal IVCs (Fig. 1). The hepatic veins drained into the right atrium. The subrenal IVC was to the left of the aorta (Fig. 2). It was shown crossing posterior to the aorta at the level of the renal veins and continuing as the azygos vein to the right of the aorta (Fig. 3). The azygos vein was coursing along the right side of the aorta in the chest (Fig. 4), and it joined the superior vena cava (SVC) (Fig. 5). The features were characteristic of an interrupted IVC and a left-sided subrenal IVC with azygos continuation. The fetus did not have any other visible anomaly. The patient had another scan at 38 weeks, at which time...
the same features were seen. A female neonate was born at term by cesarean delivery because of cephalopelvic disproportion. The postnatal period was uneventful. Sonography was done when the neonate was 43 days old. The features observed on the prenatal scans were confirmed (Fig. 6). The abdominal organs were normal.

**Case 2**
A 19-year-old gravida 1 woman was referred for sonography to rule out fetal anomalies. There was no history of consanguinity. Her menstrual age was 26 weeks. On sonography, biometric measurements corresponded to the period of amenorrhea. The fetus had a midline facial cleft in the form of a cleft of the upper lip and palate, holoprosencephaly, and a common atrioventricular valve. There was an interrupted IVC. The subrenal IVC was to the left of the aorta (Fig. 7). It continued as a hemiazygos vein along the left side of the aorta into the chest (Figs. 8 and 9). The features were characteristic of an interrupted IVC and a left-sided subrenal IVC with hemiazygos continuation. There was also a persistent umbilical vein (Fig. 8A). The parents opted for termination of the pregnancy. An autopsy was not done.

**Discussion**

Various anomalies of the IVC have been described. Knowledge of the embryogenesis of the IVC is helpful for understanding these anomalies.
The morphologic features of the IVC are the result of a dynamic process of development, regression, anastomosis, and replacement of 3 paired venous blood conduits (the posterior cardinal, supracardinal, and subcardinal veins of the primary system) starting in the fifth week. When the development of the venous system is complete, it is possible to define the embryologic origins of all its components. In particular, the SVC originates from the right superior cardinal vein. The azygos system is formed from the more cranial portion of the right (azygos) and left (hemiazygos) supracardinal veins. Blood from the subdiaphragmatic portion of the embryo is channeled into a single IVC, flowing to the paramedian right. Five embryologic segments contribute to its composition (Fig. 10). In caudal to cranial order, these segments are the iliac veins from the posterior cardinal veins, the subrenal (postrenal) segment from the right supracardinal vein, the renal segment from anastomosis between the right supracardinal and subcardinal veins, the suprarenal (prerenal) segment from the right subcardinal vein, and the hepatic segment from the hepatocardiac canal.

An interrupted IVC and a right-sided subrenal IVC with azygos continuation are the most common anomalies of the IVC. They are the result of a connection failure between the right subcardinal and right vitelline veins. Consequently, the venous blood from the caudal part of the body reaches the heart via the azygos vein and SVC.
This condition is most commonly associated with visceral heterotaxy syndromes, but, rarely, it can be an isolated condition. The prenatal sonographic features of this condition have been well reported in cases in which the condition was an isolated entity\(^4\) and part of heterotaxy syndromes\(^5\).

In an interrupted IVC, the blood of the caudal part of the body can reach the heart by other routes. The interrupted IVC can occur as a left subrenal IVC with hemiazygos continuation,\(^6\) in which the left supracardinal vein persists as the subrenal IVC instead of the right vein. Double subrenal IVCs can occur; in such cases, both IVCs join together at the renal level and continue as the azygos vein.\(^7\) There also can be double subrenal IVCs, in which the right vein continues as the azygos vein and the left continues as the hemiazygos vein.\(^8\) Postnatal diagnoses of all these types of connections have been reported. In case 1 presented here, there was agenesis of the suprarenal and hepatic segments of the IVC with a left subrenal IVC, which crossed at the renal level behind the aorta to the right, continued as an azygos vein to the right of the aorta, and drained into the SVC. The vascular anatomical structures were shown on both prenatal gray scale and color Doppler sonography at 30

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**Figure 5.** Oblique gray scale (A) and color Doppler (B) images of the fetal chest showing the azygos vein (arrowhead) joining the SVC (arrow).

**Figure 6.** A, Coronal section of the mid abdomen of the neonate from the left flank showing the left-sided subrenal IVC crossing to the right and continuing as an azygos vein. B, Coronal section of the upper abdomen from the right flank showing the azygos vein at the level of the diaphragm ascending to the right of the aorta. AO indicates aorta; AZV, azygos vein; LIVC, left subrenal IVC; LK, left kidney; and LRV, left renal vein.
weeks’ gestation. The same structures were confirmed postnatally by abdominal gray scale and color Doppler sonography. To our knowledge, the combination described in this case, namely, an interrupted IVC with a left subrenal IVC continuing as an azygos vein, has not been reported previously. In case 2, there was an interrupted IVC with a left subrenal IVC, which continued as a hemiazygos vein into the chest. The vascular anatomic structures were seen on prenatal sonography at 26 weeks’ gestation. This fetus also had a common atrioventricular valve in the heart, holoprosencephaly, a midline facial cleft, and a persistent right umbilical vein. The findings were not confirmed by autopsy. To our knowledge, this prenatal diagnosis of an interrupted IVC and left-sided subrenal IVC with hemiazygos continuation has not been reported previously, either.

Recognition of an interrupted IVC warrants a careful search for other associated anomalies, particularly cardiac anomalies. There were no other associated anomalies in case 1, but there were multiple associated anomalies in case 2. Parents have to be made aware of this condition. The dilated azygos vein is shown as a shadow on plain radiographs of the chest, but this sign should not be misinterpreted. The left subrenal IVC and the dilated azygos or hemiazygos veins should not be mistaken for enlarged lymph nodes or masses on cross-sectional imaging. Prior knowledge will help in avoiding such misinterpretations and will also be helpful when cardiac or venacaval catheterization is necessary. Finally, it is important to recognize this vascular anomaly before some surgical procedures. For example, ligation of the azygos vein during thoracotomy, which may be performed as a modification of portacaval decompression surgery, is contraindicated in patients with an interrupted IVC.

Figure 7. Transverse image of the lower abdomen of the fetus, in a longitudinal lie and breech presentation with the spine on the maternal left side, through the lower half of the kidneys, showing the subrenal IVC (arrowhead) to the left of the aorta (arrow). LK indicates left kidney.

Figure 8. Transverse gray scale (A) and color Doppler (B) images of the fetal upper abdomen showing the hemiazygos vein (arrowhead) to the left of the aorta (arrow). The persistent right umbilical vein is also shown in A. ST indicates stomach.
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Figure 9. Transverse (A) and coronal (B) gray scale and color Doppler (C) images of the fetal chest showing the hemiazygos vein (arrowhead) coursing along the left side of the aorta (arrow).

Figure 10. Final structure of the IVC and its embryologic origin. 1 indicates iliac segment, posterior cardinal veins; 2, subrenal segment, right supracardinal vein; 3, renal segment, anastomosis between the right supracardinal and subcardinal veins; 4, suprarenal segment, right subcardinal vein; and 5, hepatic segment, hepatocardiac canal.

References


